Vascular 1-1

Urticarial Vasculitis

pathogenesis, diagnosis & ttt ?

Pathogenesis:

Type III hypersensetivity Reaction

→ Circulating immune Complex → deposited on Bl Vs wall

→ Complement activation

① activates mast cells to release inflammatory Mediators

TNF-α

- ↑ ICam expression on mast Cells → eosinophil transmigration
- ↑ E selectin on endothelial Cells

② activate Complement pathway

C_3a, C_4a, C_5a

chemoattraction of neutrophils

release of proteolytic enzymes

vascular inflammation

Diagnosis ← Clinically, histopathologically, lab investigations

Clinically → painful, persistent, urticaria [- not blanch on pressure, Heal by hyperpigmentation

→ Hypocomplementemic Cases

- Major criteria
  - urticaria 7 6 Months
  - Hypocomplementemia
- Minor criteria
  - Cutaneous Vasculitis on Biopsy
  - abdominal pain
  - arthralgia
  - Glomerulonephritis
  - low C1q level

→ Normocomplementemic Cases

↓

Skin limited Ds

Q1-2 | Vasculer

lab investigation

↑ESR, hypo Complementemia, Circulating immune Comp. Complexes

Histopathology

leukocytoclastic Vasculitis

- Angiocentric Segmental inflammation
- endothelial cell Swelling
- fibrinoid necrosis of bl. Vs wall, post Capillary venules
- Cellular infiltrate around & within BL Vs wall Composed of Neutrophil & fragmented nuclei "karyorrhexis or leukocytoclasia"
- Extra vasation of erythrocytes
- Thrombosis of post Capillary venules may occur later

III

1st line of III. antihistamines.

- H_1 & H_2 antihistamines
Chlorpheniramine
- Non sedating 3rd Generation
Fexofenadine
- Ranitidine, Cimetidine
- NSAIDs indomethacine Ibuprofen
- Dapsone 100-200 mg/day
- Corticosteroids

2nd line of III

Colchicine 6mg bid
- Hydroxychloroquine
200-400 mg/day
- ~~Hydroxy~~/ Methotrexate
- azathioprine

3rd line of III

- Mycophenolate ~~Hydroxy~~/ Mofetil
- IV Ig
- Cyclosporine
Rituximab
- plasmapheresis

Henoch-Schonelin Purpura

(Anaphylactoid Purpura)

The most Comman Vasculitis in children. Primarily in male between 2-11 years.

Usually follow Streptococcal respiratory tract infection.

CIP PAPAH

- Palpable purpura (all Cases). Symmetrical hemorrhagic or purpuric lesions
- Abdominal Pain (61-67%). Colicky pain, vomiting, melena hematemesis.
- Arthralgias Arthritis - Commonly Knee Ankle.
- Renal changes - glomerulonephritis → hematuria

- Death occur Due to Renal failure

Direct immunofluorescent : IgA, C_3 & fibrinogen in thin blood vessels.

The American College of Rheumatology Criteria of HSP:-

The Presence of any 2 or more Criteria → Sensitivity 87.1%
→ Specificity 87.7%

1- palpable purpura → slightly raised hemorrhagic skin lesion
2- Bowel angina → Diffuse abdominal Pain ↑ after meal, bloody stool
3- Age ≤ 20 year at onset of Disease.
4- Vessel Biopsy → granulocytes in wall of arteriole or venule

Treatment:-

1st line tt:-

1- Supportive Care (bed rest)
2- NSAIDs for arthalgia.

2nd line.-

1- Systemic Corticosteroid
→ Treat arthritis & abdominal Pain.
↳ ↓ Duration of skin lesion.

But Not → Prevent recurrence.
↳ prevent renal Disease.

2 Dapson & Colchicine.

3- Azathioprine ± Systemic Steroids.

3rd line: - IVIg
- Rituximab
- plasma exchange.

Compare b/w
Wegner's Granulomatosis & Churg Strauss Synd.

	Wegner's Granulom	Churg-Strauss Syna
Description	described as the triad of: Granulomatous Inflammation of the UPPer & lower respiratory Tracts, Systemic necrotizing small vessel vasculitis & Pauci-immune glomerulonephritis	necrotizing granulomatous vasculitis + asthma + eosinophilia
Pathogeno	-ANCA binding to PR3 on neutrophils results in vessel damage (e.g. Pauci-immune or nonimmune complex mediated vasculitis	-Tissue inhilteration & then degranulation of eosinophils leading to tissue injury - T cells, especially th2 cells contribute to granuloma formation - ANCA-dependent activation of neutrophils

(1)

	wegner's granulomatosis	Churg Strauss Synd.
Clinically	There may be ulcerative maculopapular or erythematous lesion, Pyoderma gangrenosum over neck, thigh & buttock is not uncommon in early stage. Peri-auricular lesion destroying the ear may be one of the earliest manifestations. Death occurs from renal failure.	3 phases 1st Phase: Allergic rhinitis, nasal polyps & asthma 2nd Phase: Peripheral eosinophilia, respiratory tract infections & gastrointestinal Symp. 3rd Phase: Systemic vasculitis with granulomatous inflammation. Cut. manif during 3rd stage are palpable purpura followed by sub. cut nodules (Typically on the scalp or extremities) other systemic manifestations: mononeuritis multiplex, granulomatous inflammation of the myocardium, necrotizing G.N. Musculo-skeletal & eye involvement

2

	Wegener's granulom.	Churg Strauss Synd
lab. inv.	- Anemia, leukocytosis - ↑ ESR & CRP - c-ANCA +ve in about 80% of cases	- Eosinophilia - ANCA 60% directed against MPO, 10-15% anti-PR3 ANCA
Treatment	the standard III: systemic corticosteroids (e.g. high dose of prednisone) + cyclophosphamide - other lines: corticosteroids + Rituximab, mycophenolate mofetil, IVIg, Plasmapheresis, methotrexate, azathioprine	1st line: systemic corticosteroids 2nd line: add cyclophosphamide in cases of internal organ involvement

PROVEN LEGACY...
BETTER EFFICACY

Q 4 Vascular

Antineutrophil cytoplasmic antibodies:

- ANCA-associated vasculitis:
  The ANCA associated vasculitis are char. by
  - involvement of small to medium sized vessel,
  - The presence of ANCA
  - overlapping spectrum of organ involvement

- Pathogenesis:
  ANCA should be done in every Patient.
  Two Patterns.
  - A cytoplasmic Pattern (C-ANCA) & a Perinuclear Pattern (P-ANCA). C-ANCA, which is directed against increased Proteinase-3 (PR3), is increased in Wegner's Granulomatosis (>90%) Wegener's Granulomatosis without renal involvement (75%) & vasculitis overlap (4%-5%)
  - while (P-ANCA), which is directed against Myeloperoxidase (MPO) is found in other forms of CSVV

ANCA in vasculitis

Early

Cytokine Prim-3 leads to
↓ ANCA antigens (PR3 & MPO) becoming
expressed on the neutrophil surface

1

Q4 vasculer

2- neutrophils & endothelial cells leading to increased expression of adhesion molecules,

Mild

Circulating ANCA antibodies

Activate neutrophils to release reactive oxygen species, Pro-inflammatory mediators & chemo attractants.

late

leads to:

1- vessel wall damage

2- Recruitment of more neutrophils & inflammation

The main three ANCA- associated vasculitides:

- Microscopic Polyangiitis (MPA)
- wegner's Granulomatosis
- Churg-Strauss Syndrome

Microscopic Polyangiitis:

Pathogenesis

- Triggering Factors: Medications or Malignancy
- ANCA are thought to Play a role in the Pathogenesis

Clinical features:

constitutional symptoms (fever, weight loss, arthralgia & Myalgias)

- Cutaneous involvement: up to 70% of Patients most commonly palpable purpura, but may be erythematous patches, livedo racemosa and splinter hemorrhages

- Renal involvement (>90%), Pauci-immune crescentic necrotizing glomerulonephritis may leads to renal failure

- Pulmonary capillaritis (30-50%): Dyspnea & Pulmonary infiltrates which can result in diffuse alveolar hemorrhage

- Neurologic involvement: Peripheral neuropathy or mononeuritis multiplex

Laboratory:

Anti-MPO antibodies occur in 60% of Patients, while anti-PR3 antibodies can be seen in 30% of cases

Pathology:

- Segmental necrotizing vasculitis of the smallest blood vessels
- Vasculitis of small and/or medium sized arteries
- N.a. Granulomatous inflammation

d/q vascular

Treatment:

- Induction of remission:
- Corticosteroids (e.g. 1mg/kg/day of Prednisone) are initially used, with addition of Cyclophosphamide for Patients with significant organ involvement (e.g. renal, Pulmonary or neurologic). IV Pulse therapy can be used or
- Corticosteroids Plus rituximab
- Maintenance therapy: - Corticosteroids-sparing agents (Methotrexate, Azathioprine, MYCophenolate Mofetil, IVIG).

Q 5: Define and give Example for Grenz zone, & Leukocytoclasis?

- Grenz Zone): Narrow Zone of normal Collagen. Separate dermis from epidermis.
  Example: Granuloma fascicle- -Lepromatous leprosy- lymphocytoma.

- Leukocytoclasis: damage in and around the vessels by nuclear debris from infiltrating neutrophils - Subtypes of small vessel vasculitis.
  Example: Henoch - Schonlein purpura. (IgA vasculitis).

• Specific Tests for Wegner's granulomatosis

C-ANCA +ve in about 80% of cases

Other Tests

Anemia & leucocytosis

↑ ESR and CRP

ACR classification criteria for vasculitis Wegner's granulomatosis.

- Nasal & Oral inflammation.
- chest x Ray showing nodules, infiltrates fixed or cavities
- Microscopic haematuria or red cell casts in urine
- Granulomatous inflammation on Biopsy

Two criteria classify WG with sensitivity of 88.2% & specifity of 92.0%.

- Specific Tests for HSP

IgA, G & fibrinogen within vessel wall

the ACR classification criteria for HSP

- Palpable Purpura
- Age of onset < 20 y
- Bowel angina
- Vessel wall granulocytes on Biopsy

Two Criteria classify HSp with sensitivity of 87% & specificity of 88%

- Specific Tests for Cryoglobulinemia
  - Cryocrit: Estimation of the packed volume of precipitate after centrifugion of pre-cooled Serum ± gel electrophoresis
  - ↑ESR
  - +ve Rheumatoid factor
  - ↑ Ig level
  - Skin Biopsy.

Tests for cryoglobulins

- Assayed during clinical flares.
- on more than One occasion
- the BLood Sample should Be Kept at 37°C while Being Transported to the Laboratory

	sclerema neonatorum	subcutaneous fat necrosis of new Born
incidence →	Rare disease affecting infants during first few days of life. diffuse hardening of entire subcutaneous fat.	Rare disease affecting infants during first few days of life. fat necrosis of subcutaneous tissue.
General health of the infant →	Severely ill.	Healthy.
Sites →	Generalized.	Trunk, Buttocks, thighs
Clinically →	diffuse waxy appearance of the entire skin, Cold, sclerotic, Tight and non-Pitting induration. difficulty in Respiration and feeding.	multiple indurated non-pitting nodules and plaques → undergo spontaneous Resolution some of the nodules discharge Caseous material. hyperCalcemia may Be Associated.
histology →	• Thickning of subcutaneous Tissue • ↑ size of fat cells & presence of wide, intersecting fibrous Bands. • many fat cells are filled with fine, needle-like clefts, these clefts are occupied By crystals • Lack of inflammatory cells	• (mostly Lobular panniculitis) • foci of fat necrosis in s.c infiltrated By: lymphocytes histiocytes f.B giant cells • Ca deposits are scattered • fat cells Contain needle-shaped clefts Lie in radial fashion.
prognosis →	Poor. death occurs with in a few days.	• Excellent
treatment →	• Treatment of underlying disease • Repeated exchange Transfusion may reduce mortality	• ↑↑ of hypercalcemia By: 1- hydration 2. dietary restriction of Both Ca, vit amin D, Ca wasting diuretic (furosmide) Calcitonine & Bisphosphonate (Etidronate) · Contico steroids

Vasculan

* Q8: Management of erythema nodosum? [II]

1. Identification & treatment of the underlying disorder if identified.
A. History: of cough, dyspnea, preceding upper resp. tract infect., diarrhea, drug intake.
B. Investigations: CXR, ASOT, tuberculin test or Quanti-FERON, TB Gold test.
- Tt. of underlying infectious disease.
- Discontinue possible causative medications.

2. Treatment of the lesions themselves:
A. General measures: Bed rest, leg elantion, bandage. Legs
B. Anti-inflammatory & immuno-suppressive drugs
- Salicylates. NSAIDs.
- Potassium Iodide. (KI) Legs
- Colchicine (especially if associated Behcet's disease).
- Others: Systemic corticosteroids + hydroxychloroquine, cyclosporine, thalidomide.

N.B. ① Etanercept & infliximab have been reported to be effective in EN, but paradoxically both have been reported to produce EN as a cutaneous side effect.
② NSAIDs should be avoided in patients c IBD.

Saturated solution of Potassium iodide (SSKI)

Formulation: 1000 mg/ml

Droppers are supplied with calibrations for:

0.3 ml (300 mg) = 10 drops

0.6 ml (600 mg)

Dose:

- ✓ Adults and older children: **300 mg tid** po (starting dose: 150–300 mg tid)
- ✓ Infants and young children: 150 mg tid

Mechanism of action:

- ✓ inhibition of cell-mediated immunity
- ✓ inhibition of neutrophil chemotaxis
- ✓ suppression of neutrophil-generated oxygen intermediates

Improvement within 2 weeks

General precautions:

- ✓ SSKI should be **diluted in water or juice** to try to minimize the bitter aftertaste
- ✓ **Crystallization** may occur with **cold** temperatures, but **rewarming** and **shaking** dissolves the crystals
- ✓ Discard if solution turns **yellow–brown**

Side effects:

- **Acute:** nausea, bitter eructation, excessive salivation, urticaria, angioedema, ***cutaneous small vessel vasculitis***
- **Chronic**: enlargement of salivary and lacrimal glands, acneiform eruption, **iododerma**, **hypothyroidism**, hyperkalemia, occasionally hyperthyroidism

Vascular

Q9: Compare histopathological features of erythema nodosum & erythema induratum

EN	EI
*EN is the prototype of septal panniculitis without vasculitis. Stages:- ① Early lesions: edematous septa + mild lymphocytic infiltrates + Neutrophils. ② Miescher's microgranulomas. " Characteristic feature of EN= small collections of macrophages within septa/lobular interface, surround small cleft-like spaces). Reported in older lesions, cells have the apperance of epithelioid & multinucleated giant cells. ③ In later stages, the septa become fibrotic, partly replacing the fat lobules.	*An excisional biopsy containing adequate subcutaneous fat. 1. Mixed septal & lobular granulomatous panniculitis & neutrophilic vasculitis. 2. Caseation like necrosis may be seen.

3 stages :-
EN= lymphocytes & neutrophils → histocytes (granulomatous) → fibrotic

Neutrophilic Dermatoses

They are group of diseases characterized histologically by neutrophilic infiltrates without infectious agents [sterile pustules]

The 4 classic neutrophilic Dermatoses are.

1. Sweet's Syndrome.
2. Pyoderma gangrenosum
3. EED Erythema Elevatum Diutinum.
4. Behçet's Disease.

But there are many types of Non-infectious Dermatosis.

Epidermal.

- Pustular Psoriasis.
- IgA pemphigus.
- Drug induced acute generalized examanthematous Pustulosis.

Dermal.

- with vasculitis
  - Small vessels.
    - Henoch Schonlein Purpura.
    - urticarial vasculitis.
    - EED.
  - Medium Sized vessels
    - Polyarteritis nodosa
    - Cryoglobulinemic vasculitis.
- without vasculitis
  - DH Dermatitis herpetiform
  - Bullous SLE
  - Linear IgA bullo[us]
  - Epidermolysis bullosa acquisita
  - Sweet's Syndrome
  - Pyoderma gangrenosum
  - Behçet's Disease

Q.10 [2]

Pyoderma gangrenosum.

It is non-infectious neutrophilic dermatosis associated commonly with Systemic Disease.

Aetiopathogenesis:-

- The exact aetiopathogenesis is not fully known but there is Suggestion of:-
  - immunologic abnormality associated with Systemic Disease
  - Defect in cell-mediated immunity & humoral immunity.

clinical variants

1) classic (ulcerative) 2) Pustular PG 3) Bullous PG 4) Superficial granulomatous PG

1) Classic (ulcerative) PG:- Commonest type
   - * Symptoms:- Painful lesion with fever, malaise, arthalgia
   - * Lesions:- pustule or plaque → ulcer with Violaceous undermined edges with erythematous rim

The ulcer extends at a rapid rate & healed with Atrophic Cribriform scar especially Pretibial legs.

2) Pustular PG :- Multiple sterile pustules surrounding halo of erythema.

3) Bullous :- hemorrhagic bullae associated with hematological Dis

Q10 [3]

[4] Superficial vegetative Pg:-

- usually follow trauma (surgery)
- often solitary vegetative or ulcerative lesion

Association

1. inflammatory bowel Disease (IBD) ulcerative colitis & Crohn's
2. Arthritis.
3. Blood Disease eg leukemia.

Diagnostic Criteria

Major Criteria

1) Rapid Progression Painful necrolytic ulcer with violaceous border, an irregular undermined edge.
2) Exclusion of other causes of skin ulcer

Minor criteria

1. Presence of associated systemic Dise
2. History of pathergy or Cribriform scar.
3. Rapid response to oral corticosteroid
4. Histopathological Findings (not Specific
   → Sterile Dermal neutrophilia ±
   mixed inflamation ±
   lymphocytic vasculitis & exclu
   infective cause.

Diagnosis Need Both All major + at least 2 minor crite

Evaluation

- GIT → Stool analysis, radiography, liver function test
- Blood Study → CBC, ANA, ANCA antibodies, chest Xray a

Treatment

Aim → Control underlying Disease.
↳ ↓ inflammatory process of wound & induce healing.

1) general measure (Ulcer care):-
- Compression.
- limb elevation.
- Skin grafting.
- flap repairs.
- Dressing.

2) Adjuvant therapy:- intralesional Corticosteroid.
- Super potent topical corticosteroids.
- oral Antibiotic.
- Dapson (50-150mg tunic Da

3) Severe Case :- Prednisone 60-120mg daily.
Cyclosporine 2.5-5mg/kg/day.
- IVIG 2-3gm/Kg IV slowly.

4) Treatment of associated Disease.

q 11

Sweet syndrome

Acute Febrile neutrophilic Dermatosis

Epidemiology: Age → Any age Female > male (4:1)

Etiopathogenesis: unknown.

- Genetic: Abnormality of chromosom 3q
  HLA - B54
- Hypersensitivity reaction
  - Ig and Complement - mediated activation and mobilization of Neutrophilis.
  - Antigen or super Antigen - induced T-cell dependent cellular immune Reaction
- Immune cytokine dysregulation (local or systemic)

Certain cytokines and chemokines may help in initiation and propagation of inflammatory process of sweet syndrome

eg IL 1, IFNγ - GM-CSF, G-CSF
granulocyte-macrophage colony stimulating Factor | Granulocyte-colony st.F

Clinical Features:

- ⊕ FHMA
- ⊕ Cutaneous manifestations: Acute onset of Asymmetrically distributed Bright red to purple, painful tender, warm, sharply demarcated plaques which have mamillated surface the Lesion may be annular, arcute, acneiform.
- Site: Face, trunk, upper extremities
- oral and genital Lesion are rare but may be occur.
- +ve pathergy test: development of specific skin Lesion at the site of minor truama or injury (needle) it's positive in 3% only.
- Resolve spontaneously within 5-12 weeks but may recurrent in 30%

⊛ Extra cutaneous manifestations:

→ Serum sickness symptoms:- Myalgia, Malaise, Headache, N
Musculo-Skeletal → arthralgia, arthritis of elbow, knees
Ocular → iritis, Conjunctivitis, iridocyclitis.
Respiratory → Cough, dyspnea, pleurisy, P. effusion
Renal → Haematuria, Proteinuria, Acute Renal Failure.
Meningeal → Meningitis (Aseptic), encephalitis.
GIT → Hepatitis, Pancreatitis, GIT upsets.

D. Criteria

① Major :→ Skin lesion + Histo pathological Finidings
② Minor :→ ⊙ Preceded by Fever or infection
⊙ Fever > 38°c
⊙ Leucocytosis
⊙ Associated with extra cutaneous manifestation
⊙ Dramatic Response to systemic steroid or potassium Iodide Not to Systemic Antibiotics

How to diagnose → 2 major + 2 minor

Histopathology :→

① upper and mid-dermal perivasular diffuse Neutrophilic in filtrate
② Papillary dermal edema and ~~RBC// extravasation~~
③ Leukocytoclasia (nuclear dust) with endothelial swelling, vasodilatation, extravasation of erythrocyte But without fibrinoid necrosis DD of vasculitis.

Lab finindings :-

⊙ ↑ ESR
⊙ Neutrophilia
⊙ ↑ CRP
⊙ Leucocytosis
⊙ Antineutrophil cytoplasmic Abocs (ANCA)

Association with sweet's syndrome

[1] Malignancy:

Leukemias - Breast cancer - ovarian cancer
Endometrial Cancer, Vaginal Cancer
Rectal Cancer, Prostatic cancer, testicular Cancer.

[2] Inflammation:-

GIT ulcerative colitis, Crohn's disease, Cirrhosis
sub acute thyroiditis, Behcet syndrome,
Rheumatoid arthritis, pyoderma gangrenosum.

[3] Infection:

(S) Staph, strept, salmonella.
(H) Hepatitis, HIV, histoplasmosis
Meningitis, TB.

[4] Others

Drugs, Pregnancy, Renal stone, Immunization, Photoinduction

Treatment

- Antibiotics is ineffective until if associated with Bact. infection
- Topical Rx in Few or localized Lesion
  Topical CST or Calcineurin inhibitors
- Systemic Rx
  Prednisolone (0.5 - 1.0) mg/kg/day
  for 4 - 6 weeks.

Others Drugs → K. iodide 9000 mg/day / Colchicine
Dapsone (100 - 200 mg/day) / Cyclosporine (5 - 10 mg/day) ↓ IL1 production
Indomethacin 50

Q 12-1

(12) - Mon diagnostic criteria for Henoch-schonelin purpura (HSP)

① palpable purpura

② Abdominal pain

③ Arthralgia and arthritis

④ Renal changes: hematuria

⑤ Age ≤ 20 yrs at disease onset

* Behcet disease

① Recurrent aphthous stomatitis

② Recurrent painful genital ulcers.

Q 12-2

Subject: Day: Date:

③ Ocular lesions (70-85%)
e.g: Keratitis, optic neuritis

④ CNS lesions, e.g: cranial nerve palsies, meningoencephalitis

⑤ other cutaneous lesions:-
"pathergy" occurs 40-90% d.pt.
, it's Diagnostic

(Q13) Pathogensis & managment of Behcet's disease

* Behcet disease is a chronic multisystem disease characterized by oral & genital aphthae, cutaneous lesions, ocular, gastrointestinal & neurologic manifestations

* must be distinguished from complex aphthosis which is characterized by the presence of almost constant multiple ≥3 oral, or oral & genital aphtae in the absence of systemic manifestations

"Pathogensis"

(1) Genetic background: suggested by its association with HLA-B51

(2) Infections: HSV, hepatitis C virus parvovirus B19 and more recently streptococcal strains these infections may potentially trigger an immunoregulatory defect in the genitically predisposed

(3) Autoimmune responses:

(a) circulating immune complexes: patients with Behcet's have higher number of circulating immune complexes

(b) Role of neutrophils: There's enhanced neutrophil chemotaxis, activation & production of Ros & lysosomal enzymes → tissue injury

(c) Role of cytokines: & the levels of cytokines IL-12 (promotes Th1 response) IL8 → neutrophil chemoattractant collect with disease activity

④ Clonal expansion of autoreactive T-cells that recognized a peptide derived from heat shock protein 60

⑤ High prevalence of procoagulant mutations and increased platelet activation has also been suggested as an explanation for the thrombotic component of the disease

* Diagnosis of Behcat's disease

1- Recurrent oral ulceration: minor aphthous, major aphthous, herpitiform ulceration — received at least 3 times in one 12 month period.

② Plus two of the following criteria

2- Recurrent genital ulceration: aphtous ulceration or scarring

3- eye lesions → Anterior uveitis, Retinal vasculitis

4- Skin lesions → erythema nodosum, pseudofolliculitis, papulopustular lesion, acneform nodules in post adolescent patients not receiving corticosteroid th

⑤ +ve pathergy test

Page : Date :

* treatment of Behcat's disease

[1] Mucocutaneous disease :-
- intralesional corticosteroids
- colchicine
- Dapsone
- Combination of oral colchicine + Dapsone
- topical corticosteroids
- viscous lidocaine, topical sucralfate

[2] Severe mucocutaneous disease
- Thalidomide 50-150 mg po nightly
- Methotrexate 2.5-5 mg po or IM weekly
- prednisone 40-80 mg po
- Interferon α 2a
- TNF α inhibitors e.g. ~~ant~~ etanercept, infliximab)

[3] systemic disease
- prednisone 60-120 mg po daily
- azathioprine 50-100 mg po twice daily
- Cyclosporine
- IVIg
- TNF α inhibitors

Q 14-1

(B) Eye manifestation of Behcet disease

- Anterior uveitis, Posterior uveitis
- Cells in vitreous on slit lamp examination, or retinal vasculitis observed by ophthalmologist
- ~~Keratitis, optic neuritis,~~ uveitis and may be blindness.

Good luck

1 Treatment

Q 15 Therapeutic option for Pyoderma gangrenosum?

There is neither specific nor effective therapy for PG.

- **Goals of therapy:** to reduce the inflammatory process of the wound, promote healing, reduce pain and to control the underlying disease (especially leukemias and IBD).
- **Choice of therapy:** the therapeutic approach depends on the number, size and depth of the lesions, the rate of expansion and appearance of new lesions, the associated disorder, the medical status of the patient, and the risk and patient tolerance of prolonged therapy.
- **Lines of treatment:**

(1) General measure (ulcer care):

- Compression
- Limb elevation
- Hyperbaric oxygen
- Low-pressure / Bio-occlusive dressings
- Skin grafting, flap repairs

2nd Q15 th of PG

2

(2) Mild disease / adjuvant therapy:

- Superpotent topical / intralesional corticosteroids
- Topical tacrolimus
- Oral antibiotics (e.g. sulfonamides, minocycline)
- Colchicine (0.6 mg thrice daily) and/or Dapsone (50–150 mg thrice daily)
- Clofazimine 100–400 mg daily
- Other (e.g. oral potassium iodide, intralesional cyclosporine, topical cromolyn sodium, nicotine patch or cream)

(3) More severe disease:

- **Prednisone 60–120 mg daily**
- Methylprednisolone 1 g daily for 3–5 days (IV pulse)
- **Cyclosporine 2.5–5 mg/kg daily**
- Thalidomide 50–150 mg every night
- TNF-α inhibitors: Infliximab, adalimumab, etanercept
- Methotrexate 2.5–25 mg orally or IM weekly
- Azathioprine 50–100 mg twice daily
- Mycophenolate mofetil 1–1.5 g orally twice daily
- Cyclophosphamide
- IVIG 2–3 g/kg IV (given over 2–5 consecutive days)
- Plasmapheresis

(4) Treatment of the underlying disease (if present):

- Total colectomy (for severe chronic ulcerative colitis)

Q 16. Histopathology of Degos disease?
(malignant atrophic papulosis).

H/P: Endothelial swelling and thrombosis of small arteries & absent inflammation of vessel wall - a wedge shape area of necrosis of the dermis in which Collagen appears homogenous and devoid of nuclei - much deposits are found in the area of necrosis - the stratum malpighii markedly atrophic.